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PAEDIATRIC RESPIRATORY MEDICINE

Jeremy Hull
Julian Forton
Anne Thompson

SECOND EDITION

PAEDIATRICS

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Oxford Specialist Handbooks in Paediatrics

Paediatric Respiratory Medicine

Second Edition

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Preface to the Second Edition

In writing this new edition, we have gone through each chapter, bringing sections up to date, where needed, and including key new references. Following the publication of the Global Lung Initiative prediction equations for spirometric lung volumes, the section on lung function normal values has been removed. Otherwise, the structure of the book is unchanged. The aim of the handbook is to assist clinicians in the day-to-day care of children with respiratory disease. For rarer conditions, there is perhaps more detail than the reader might expect—this reflects our view that, when faced with a child with a rare condition, a bit of background reading is required! The topics covered in the book include all those required for the European Respiratory Society Hermes exam in paediatric respiratory medicine, and so this handbook is a useful text for those considering taking that qualification.

JH, JF, AHT 2015

Preface to the First Edition

In writing this handbook, our aim has been to provide a practical guide to paediatric respiratory medicine that will be a useful first point of reference for paediatricians faced with children with acute or chronic respiratory problems.

The book has been written for use by general paediatricians and by specialists in paediatric respiratory medicine at both consultant and trainee level. It deals with common problems seen by all paediatricians and rarer conditions more often seen at specialist centres, but managed in collaboration with general paediatricians. The information is in a readily accessible format, with extensive use of bullet points. The focus is on clinical presentation, diagnosis, and management of respiratory problems. There is less emphasis on background information, such as epidemiology and pathogenesis, but this is sufficient where necessary to provide insight into clinical presentation and management, or where this information would be helpful for parents.

The book is divided into four parts. Part 1 provides a practical approach to acute and non-acute clinical problems. Part 2, the bulk of the book, provides detailed information about common and not-so-common clinical conditions. Part 3 provides useful information on supportive care, including, for example, use of non-invasive ventilation and the care of a child with a tracheostomy. Part 4 gives details on how to perform several practical procedures, such as ciliary brush biopsy, flexible bronchoscopy, and inserting a chest drain. Finally, the appendices provide information on lung function testing and tables of age-corrected normal values for several respiratory parameters.

The book has been written by two consultants who work in a tertiary respiratory unit and by a respiratory trainee. This combination of authors has provided the experience necessary to deal with topics where there is an absence of published evidence and to present the information in a format that both consultants and trainees will find useful.

JH, JF, and AHT July 2007

Foreword to the First Edition

So why would anyone want to buy a book in the 21st century, when a profusion of information is available at the touch of a computer key? Cough in children yields more than 6500 references on a PubMed search, so why would anyone want to read a 5 page section on the same subject in this book? One of the most misquoted phrases in English poetry, 'a little *learning* is a dangerous thing' could now be adapted as 'a lot of PubMed is a dangerous thing', and unless the searcher has a sound grounding in the subject searched, it is likely to lead to confusion in mind, an unbalanced perspective, and ultimately, inappropriate management of children. A view of the woods is an essential prelude to a detailed study of an individual tree.

And that is where this book will be so valuable to all in the field of paediatric respiratory medicine, from the raw young tyro to the elderly professorial dodderer, and all stages in between. The reader will find a clear account of the subject, from both a problem-based and a disease-based approach. It gives a commonsense overview of all the important topics in the field, with crisp tables and bullet points, written in clear English. There are a few, up-to-date papers, reviews and websites as a basis for further learning, and the authors have fully achieved their aim, of writing a practical handbook in line with the long tradition of the *Oxford* series. The trainee can rapidly acquire a good grasp of the subject, and can then safely dive into PubMed for more advanced studies, particularly of pathophysiology. Few if any of the allegedly trained will read this without finding something to learn, or some new idea not previously thought of, to try when next a problem arises. Few will agree with absolutely every statement, but that is inevitable and part of the intrinsic beauty of the subject—and in any event, medicine is learned by doing, and cannot be learned solely from books, whatever the views of those currently changing medical training with all the natural talent of a hippopotamus playing the piccolo.

So in summary, who can benefit from this book, and how? The trainee will certainly not outgrow it—even the most experienced paediatrician, seeing a child with an uncommon condition, or preparing a teaching session on a common one, will benefit from taking a surreptitious peek at the relevant section here, to ensure nothing has been forgotten. For example, I would challenge the reader to list the totality of the associated conditions which need to be detected in a baby with a PHOX2b mutation before turning to Chapter 26. Review copies of books come into three categories: 'throw away', 'give away' and 'chain it to the wall'—this Handbook is definitely in the last category. Departmental thieves, hands off!

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We gratefully acknowledge the skill and patience of Professor Sir David Hull who drew most of the figures.

Contents

Symbols and abbreviations

±	plus/minus
=	equal to
>	more than
<	less than
≥	equal to or more than
≤	equal to or less than
%	per cent
β	beta
γ	gamma
£	pound sterling
°C	degree Celsius
®	registered
™	trademark
AAD	adaptive aerosol delivery
AAT	alpha-1 antitrypsin
ABG	arterial blood gas
ABPA	allergic bronchopulmonary aspergillosis
ACBT	active cycle of breathing technique
ACD	alveolar capillary dysplasia
ACE	angiotensin-converting enzyme
Ach	acetylcholine
AChE	acetylcholinesterase
AChR	acetylcholine receptor
ACM	Arnold–Chiari malformation
ACS	acute chest syndrome
ADA	adenosine deaminase
ADHD	attention-deficit/hyperactivity disorder
AHI	apnoea hypopnoea index
AIDS	acquired immune deficiency syndrome
ALL	acute lymphoblastic leukaemia
ALT	alanine aminotransferase
ALTE	apparent life-threatening event
ANA	antinuclear antibody
ANCA	antineutrophil cytoplasmic antibody
ANSD	autonomic nervous system dysregulation
APTT	activated partial thromboplastin time

ARDS	acute respiratory distress syndrome
ART	antiretroviral therapy
AS	adolescent scoliosis
ASD	atrial septal defect
AST	aspartate aminotransferase
AT	ataxia telangiectasia
atm	atmosphere
ATP	adenosine triphosphate
ATS	American Thoracic Society
AXR	abdominal X-ray
BAL	bronchoalveolar lavage
BCG	bacille Calmette–Guérin
BD	Behçet's disease
BE	base excess
BHL	bilateral hilar lymphadenopathy
BIPAP	bilevel positive airway pressure
BMD	bone mineral density; Becker muscular dystrophy
BMI	body mass index
BO	bronchiolitis obliterans
BOOP	bronchiolitis obliterans and organizing pneumonia
BOS	bronchiolitis obliterans syndrome
BPD	bronchopulmonary dysplasia
bpm	beats per minute
BSLT	bilateral sequential lung transplantation
BTS	British Thoracic Society
Ca	calcium
c-ANCA	cytoplasmic antineutrophil cytoplasmic antibody
CBF	ciliary beat frequency
CCAM	congenital cystic adenomatoid malformation
CCHS	congenital central hypoventilation syndrome
CDC	Centers for Disease Control and Prevention
CEA	carcinoembryonic antigen
CF	cystic fibrosis
CFC	chlorofluorocarbon
CFRD	cystic fibrosis-related diabetes
CGD	chronic granulomatous disease
CHARGE	coloboma, heart defects, atresia choanae, retarded growth and development, genital hypoplasia, ear abnormalities
ChAT	choline acetyl transferase
chILD	children's interstitial and diffuse lung disease

CIP	chronic interstitial pneumonitis
CLAD	chronic lung allograft dysfunction
CLD	chronic lung disease (of prematurity)
CLE	congenital lobar emphysema
cmH_2O	centimetre of water
CMI	cell-mediated immunity
CMV	cytomegalovirus
CNS	central nervous system
CO	carbon monoxide
CO_2	carbon dioxide
COHb	carboxyhaemoglobin
CPAM	congenital pulmonary airway malformation
CPAP	continuous positive airway pressure
CPF	cough peak flow
CRP	C-reactive protein
CSF	cerebrospinal fluid
CT	computed tomography
CVID	common variable immune deficiency
CXR	chest X-ray
DEXA	dual-energy X-ray absorptiometry
DIOS	distal intestinal obstruction syndrome
DIP	desquamative interstitial pneumonitis
dL	decilitre
DMD	Duchenne muscular dystrophy
DNA	deoxyribonucleic acid
2,3-DPG	2,3-diphosphoglycerate
DPI	dry powder inhaler
DRV	daily recommended value
dsDNA	double-stranded deoxyribonucleic acid
EBV	Epstein–Barr virus
ECG	electrocardiogram
ECMO	extracorporeal membrane oxygenation
EEG	electroencephalography
ELISA	enzyme-linked immunosorbent assay
ELS	extralobar sequestration
EM	electron microscopy
EMG	electromyography
ENaC	epithelial sodium channel
ENT	ear, nose, and throat
ERS	European Respiratory Society

ESR	erythrocyte sedimentation rate
ETT	endotracheal tube
F	French
FBC	full blood count
FEES	fibreoptic endoscopic evaluation of swallowing
FEF	forced expiratory flow
FENO	fractional exhaled nitric oxide
FEV_1	forced expiratory volume in 1 second
FFP	fresh frozen plasma
FGFR	fibroblast growth factor receptor
FiO_2	fraction of inspired oxygen
FISH	fluorescent in situ hybridization
FRC	functional residual capacity
ft	foot/feet
FVC	forced vital capacity
g	gram
GAG	glucosaminoglycan
GAS	group A Streptococcus
GBS	Guillain–Barré syndrome
GCR	glucocorticoid receptor
G-CSF	granulocyte colony-stimulating factor
GD	Gaucher's disease
GH	growth hormone
GI	gastrointestinal
GINA	Global Initiative for Asthma
GLI	Global Lung Initiative
GM-CSF	granulocyte macrophage colony-stimulating factor
GOR	gastro-oesophageal reflux
GORD	gastro-oesophageal reflux disease
GP	general practitioner
GTT	glucose tolerance test
GVHD	graft-versus-host disease
h	hour
HAART	highly active antiretroviral therapy
Hb	haemoglobin
HBoV	human bocavirus
HCG	human chorionic gonadotrophin
HDU	high dependency unit
HEPA	high-efficiency particle arrest
HFCWO	high-frequency chest wall oscillation

Hib	*Haemophilus influenzae* type b
HIV	human immunodeficiency virus
HLA	human leucocyte antigen
HMP	human metapneumovirus
HP	hypersensitivity pneumonitis
HPV	human papillomavirus
HRCT	high-resolution computed tomography
HVA	homovanillic acid
IC	inspiratory capacity
ICU	intensive care unit
Ig	immunoglobulin
IGF	insulin growth factor
IGT	interferon-gamma test
IL	interleukin
ILD	interstitial lung disease
ILS	intralobar sequestration
IM	intramuscular
IPH	idiopathic pulmonary haemosiderosis
IQ	intelligence quotient
IRT	immune-reactive trypsin
ITP	idiopathic thrombocytopenic purpura
IU	international unit
IUGR	intrauterine growth retardation
IV	intravenous
IVC	inferior vena cava
IVIg	intravenous immunoglobulin
JATD	Jeune asphyxiating thoracic dystrophy
JCVI	Joint Committee on Vaccination and Immunisation
JDM	juvenile dermatomyositis
JIA	juvenile idiopathic arthritis
JORRP	juvenile onset recurrent respiratory papillomatosis
K	potassium
kcal	kilocalorie
kCO	transfer factor
kg	kilogram
kPa	kilopascal
L	litre
LABA	long-acting beta-agonist
LAM	lymphangioleiomyomatosis
LCFA	long-chain fatty acid

LCH	Langerhans cell histiocytosis
LCI	lung clearance index
LCT	long-chain triglyceride
LDH	lactate dehydrogenase
LIP	lymphoid interstitial pneumonitis
LOW	late-onset wheeze
LRTI	lower respiratory tract infection
LTEC	laryngotracheo-oesophageal cleft
LTOT	long-term oxygen therapy
m	metre
MBC	maximum breathing capacity
MCFA	medium-chain fatty acid
MCT	medium-chain triglyceride
MDI	metered-dose inhaler
MDR	multidrug resistance
mEq	milli equivalent
mg	milligram
Mg	magnesium
MG	myasthenia gravis
min	minute
mL	millilitre
MLS	middle lobe syndrome
mm	millimetre
mmHg	millimetre of mercury
mmol	millimole
mph	mile per hour
MPS	mucopolysaccharidoses
MRA	magnetic resonance angiography
MRI	magnetic resonance imaging
mRNA	messenger ribonucleic acid
MRSA	meticillin-resistant Staphylococcus aureus
MSSA	meticillin-sensitive Staphylococcus aureus
MTB	Mycobacterium tuberculosis
mTOR	mammalian target of rapamycin
Na	sodium
NBT	nitroblue tetrazolium
NEB	neuroepithelial body
NEHI	neuroendocrine hyperplasia of infancy
NG	nasogastric
NHL	non-Hodgkin's lymphoma

NHS	National Health Service
NICE	National Institute for Health and Care Excellence
NIH	National Institutes of Health
NIPPV	non-invasive positive pressure ventilation
NIV	non-invasive ventilation
NK	natural killer
nmol	nanomole
NO	nitric oxide
NPA	nasopharyngeal aspirate
NPARM	non-polyalanine repeat mutation
NPD	Niemann–Pick disease
NREM	non-rapid eye movement
NSAID	non-steroidal anti-inflammatory drug
NSIP	non-specific interstitial pneumonitis
NTM	non-tuberculous mycobacteria
OGTT	oral glucose tolerance test
OP	organizing pneumonia
OSA	obstructive sleep apnoea
PAA	pulmonary artery aneurysm
p-ANCA	perinuclear antineutrophil cytoplasmic antibody
PAP	pulmonary alveolar proteinosis
PARM	polyalanine repeat expansion mutation
PAS	periodic acid–Schiff
PBMC	peripheral blood mononuclear cell
PC	pectus carinatum
PCD	primary ciliary dyskinesia
PCP	pneumocystis pneumonia
PCR	polymerase chain reaction
PCV	pneumococcal conjugate vaccine
PE	pectus excavatum; pulmonary embolism
PEEP	positive end-expiratory pressure
PEFR	peak expiratory flow rate
PEP	positive expiratory pressure
PIE	pulmonary infiltrates with eosinophilia
PIG	pulmonary interstitial glycogenesis
PIV	parainfluenza virus
PL	pulmonary lymphangiectasia
PLMD	periodic limb movement disorder
PLMS	periodic limb movement in sleep
PMA	post-menstrual age